

Teaching Video NeuroImages: Cautious walking gait in siblings with hereditary hyperekplexia

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A 48-year-old man and his 41-year-old sister complained of pathogenic startles since birth. They carried a homozygous deletion mutation in *GLRA1*: c.754delC (p. L252X). They felt stiff in 4 limbs with expressionless face when they were alarmed by unexpected touch and sound stimulation, startled, and fell down with stiffed limbs since walking independently. Therefore they adopted a cautious walking gait as a subconscious self-protection (videos 1 and 2).

With clonazepam treatment, pathologic cautious walking gait can be dramatically improved. Cautious walking gait is an interesting and easily observable manifestation of hereditary hyperekplexia.^{1,2}

Disclosure

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